https://doi.org/10.30702/ujcvs/23.31(02)/PB028-1624
UDC 616.126.42:616.126.6]-089

Volodymyr V. Popov[1], Ph.D., Doctor of Medical Science, Professor, Head of the Department of Surgical Treatment of Acquired Heart Diseases, https://orcid.org/0000-0002-2851-5589

Oleksandr O. Bolshak[1], Ph.D., Senior Research Fellow of the Department of Surgical Treatment of Acquired Heart Diseases, https://orcid.org/0000-0002-6089-9594

Valery Zh. Boukarim[1], Junior Researcher of the Department of Surgical Treatment of Acquired Heart Diseases, https://orcid.org/0000-0002-3895-1894

Olena V. Khoroshkovata[1], Ph.D., Cardiologist of the Department of Surgical Treatment of Acquired Heart Diseases, https://orcid.org/0000-0002-5857-4643

Oleksii M. Gurtovenko[1], Anesthesiologist of the Department of Surgical Treatment of Acquired Heart Diseases, https://orcid.org/0000-0002-3163-4327

Evgen V. **Novikov**[2], Ph.D., Assistant of the Department of Propaedeutics of Internal Medicine, Radiation Diagnostics and Therapy, https://orcid.org/0000-0002-5864-0604

[1]National Amosov Institute of Cardiovascular Surgery of the National Academy of Medical Sciences of Ukraine, Kyiv, Ukraine

[2]Zaporizhzhia State Medical University, Zaporizhzhia, Ukraine

Surgical Reconstruction of Mitral-Tricuspid Insufficiency Combined with Biatriomegaly in Marfan Syndrome

Abstract

The aim. To analyze the results of introduction of complex reconstruction of the left and right parts of the heart in Marfan syndrome combined with mitral-tricuspid insufficiency, biatriomegaly and left ventriculomegaly.

Materials and methods. As an example of this pathology, we present our observation. Female patient T., 24 years old, was examined and treated from 7/27/2022 to 8/17/2022 at the Department of Surgical Treatment of Acquired Heart Diseases of the National Amosov Institute of Cardiovascular Surgery of the National Academy of Medical Sciences of Ukraine with a diagnosis: Marfan syndrome. Stage IV mitral-tricuspid insufficiency. Biatriomegaly. High pulmonary hypertension. Permanent form of atrial fibrillation (for 10 years, since 2011). IIB heart failure with left ventriculomegaly with reduced left ventricular ejection fraction. NYHA functional class IV+. The patient underwent surgery in the following volume: mitral valve replacement with full preservation of the valve apparatus + tricuspid valve plasty with the imposition of a support ring + triangular plasty of the left atrium with its partial resection + resection of the right atrium.

Results. Within 4 months after surgery, echocardiographic parameters showed significant decrease in the volume of the left ventricle, the diameters of both atria. In addition, there was a significant decrease in the level of N-terminal pro-brain natriuretic peptide and manifestations of heart failure.

Conclusion. Taking into account the initial serious condition of the patient with Marfan syndrome and advanced mitral-tricuspid heart disease, left ventriculomegaly with a reduced left ventricular ejection fraction, biatriomegaly (left atrium 7.2 cm, right atrium 6.5 cm), permanent form of atrial fibrillation, complex reconstruction of the left and right parts of the heart leads to an improvement in the functional state of the myocardium.

Keywords*: cardiac surgery, atrial fibrillation, mitral-tricuspid malformation, left ventriculomegaly, cardiopulmonary bypass, sinus of Valsalva dilatation.*

Background. Marfan syndrome is a clinically significant risk factor for surgical treatment of patients with mitral-tricuspid heart disease. Concomitant left atriomegaly often leads to compression of the left ventricle (LV), bronchi, resulting in significant respiratory disorders and heart failure (HF). Significant left atrial dilatation prevents the restoration of sinus rhythm and increases the risk of thromboembolic complications [1, 2, 3]. The constant form of atrial fibrillation (AF) significantly affects the contractility of the LV.

Left ventriculomegaly is an additional significant risk factor for surgery in Marfan syndrome and correction of mitral-tricuspid defect [4, 5].

Due to connective tissue dysplasia, complete excision of the valvular apparatus and mitral valve replacement in Marfan syndrome is an unacceptable procedure, since this can lead to rupture of the LV wall, and most importantly, to a decrease in its contractility.

Despite the neglect of the patient's condition, there is an expediency of a comprehensive reconstruction of the left and right parts of the heart.

The emerging clinical contradictions between the pronounced valvular pathology of the mitral-tricuspid valve are dissonant with the absence of a pronounced pathology on the aortic valve, as well as the ascending aorta. Severe HF combined with tachyforms of AF actually makes waiting for drug therapy fruitless and dictates the expediency of performing surgical correction of only the valvular component of the pathology combined with atrial dilation, while at the same time postponing to an indefinite future a possible problem with the condition of the tissues of the ascending aorta in the period 35-40 years of the patient's life. Limiting the time of anoxia allows only a full-fledged intervention, with the exception of the ascending aorta. The problem of arrhythmia is taken out in the projection of the early postoperative period (from six months to 1 year).

The presence of risk factors (family history of Marfan syndrome, increase in the size of ascending aorta > 2-3 mm/year, need for aortic valve replacement), surgery should be considered for the ascending aorta diameter ≥ 45 mm [6].

The aim. To analyze the results of introduction of complex reconstruction of the left and right parts of the heart in Marfan syndrome combined with mitral-tricuspid insufficiency, biatriomegaly and left ventriculomegaly.

Materials and methods. Female patient T., 24 years old, was treated from 7/27/2022 to 8/17/2022 at the Department of Surgical Treatment of Acquired Heart Diseases of the National Amosov Institute of Cardiovascular Surgery of the National Academy of Medical Sciences of Ukraine with a diagnosis: Marfan syndrome. Stage IV mitral-tricuspid insufficiency. Biatriomegaly. Left ventriculomegaly. Permanent form of AF (for 10 years, since 2011). IIB HF with reduced left ventricular ejection fraction (LVEF). NYHA functional class IV+.

The patient complained of shortness of breath, swelling of the lower extremities, interruptions in the work of the heart. According to the patient, over the past year she has lost 8 kg. There was significant hypotrophy. With a height of 175 cm, body weight was 52 kg (deficiency was 13 kg). Significant asthenia. Visual apparatus was not damaged, however, changes in the limbs were noted (Fig. 1).

The patient's clinical condition was severe based on N-terminal pro-brain natriuretic peptide (NT-proBNP) level (11 356 pg/ml, with normal range from 0 to 220.0), which was 51.6 times higher than normal. In this regard, during the 7 days of preoperative examination, the patient received Uperio at a dose of 100 mg twice daily, as well as dexamethasone intravenously 8.0 mg once daily in the morning, digoxin 0.005 g per day. Biochemical and clinical parameters were unchanged. Hemoglobin 141 g/l, red blood cells $4.5x10^{12}$/l.

According to the echocardiography before surgery (Table 1), critical mitral insufficiency was identified due to the prolapse of both valves of myxomatous origin, which is characteristic of Marfan syndrome (Fig. 2), significant tricuspid insufficiency, biatriomegaly, left ventriculomegaly (double and more increase).

Percent deviation 7 and 14 days after the operation was calculated based on preoperative values.

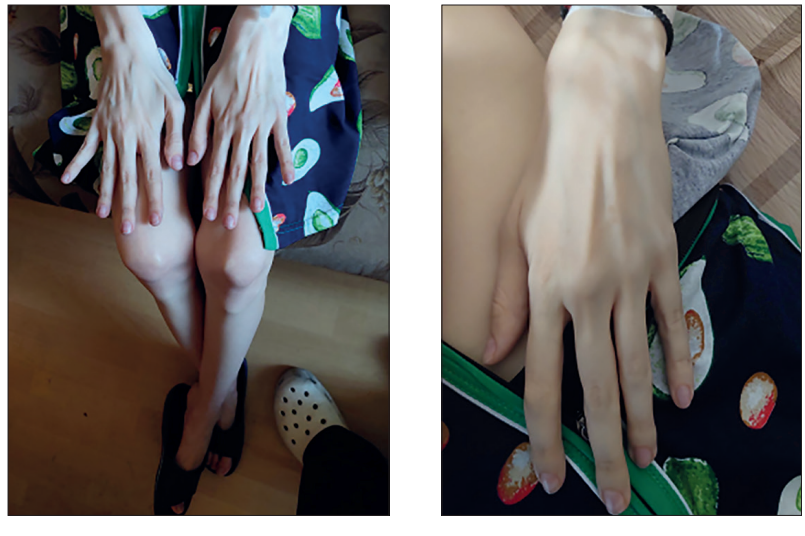

Fig. 1. *The patient's limbs*

Table 1

Echocardiographic parameters at the hospital stage

Parameters (units)	Before the surgery (7/27/2022)	7 days after surgery (8/10/2022)	14 days after surgery (8/17/2022)
Mitral valve	Myxomatous degeneration of the leaflets, prolapse of both valves, reverse leakage ++++	Prosthesis, reverse leakage minimum (+), pressure gradient 7 mm Hg	Prosthesis, reverse leakage minimum (+), pressure gradient 7 mm Hg
Tricuspid valve	Prolapse, reverse leakage pronounced +++	Condition after tricuspid valve plasty, reverse leakage +	Condition after tricuspid valve plasty, reverse leakage small (+), pressure gradient 3 mm Hg
Aortic valve	Tricuspid, compacted, Δp max 10 mm Hg, reverse leakage minimal, diameter of the mouth of the aorta 2.2 cm, sinus of Valsalva diameter 3.9 cm, ascending aorta 3.0 cm		Diameter of the ascending aorta 3.1 cm, sinus of Valsalva diameter 3.9 cm
EDVI (ml/m^2)	233.12	139.37 (-40.2%)	110.62 (-52.5%)
LV EDV (ml)	373	223 (-40.2%)	177 (-52.5%)
ESVI (ml/m^2)	109.37	75 (-31.4%)	55 (-49.7%)
LV ESV (ml)	175	120 (-31.4%)	88 (-49.7%)
LV SVI (ml/m^2)	123.75	64.37 (-48.0%)	55.62 (-55.0%)
LV SV (ml)	198	103 (-48.0%)	89 (-55.0%)
LVEF	0.53	0.46 (-13.2%)	0.5 (-5.6%)
LA diameter (cm)	7.2	5.1 (-29.1%)	5.1 (-29.1%)
SPAP (mm Hg)	40	38 (-5.0%)	38 (-5.0%)
RA diameter (cm)	6.5	3.9 (-40.0%)	3.7 (-43.1%)

EDVI, end-diastolic volume index; ESVI, end-systolic volume index; LA, left atrium; LV EDV, left ventricular end-diastolic volume; LV ESV, left ventricular end-systolic volume; LV SV, left ventricular systolic volume; LV SVI, left ventricular systolic volume index; RA, right atrium; SPAP, systolic pulmonary artery pressure.

Chest X-ray revealed cardiomegaly and small right-sided hydrothorax (Fig. 3).

According to the results of coronarography (7/29/2022), atherosclerotic lesions of the coronary arteries were not detected.

EuroSCORE II score was 3.9%.

The operation was performed on 8/2/2022, the volume of the operation was as follows: mitral valve replacement with full preservation of the valve apparatus

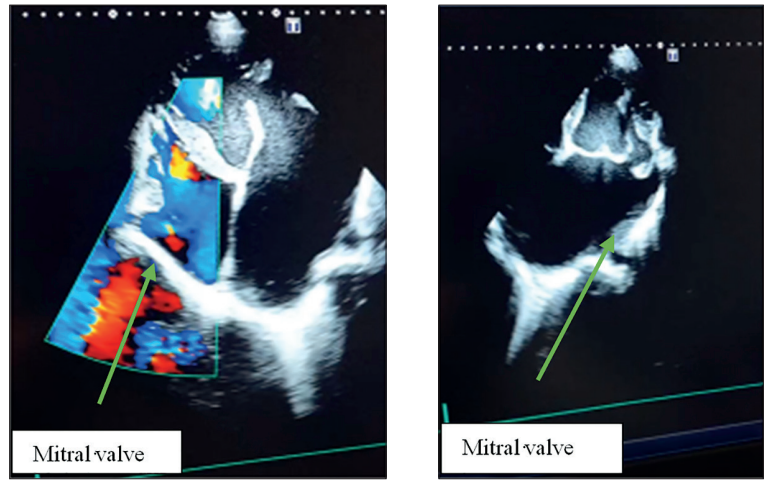

Fig. 2. *Myxomatous changes on the mitral valve characteristic of Marfan syndrome*

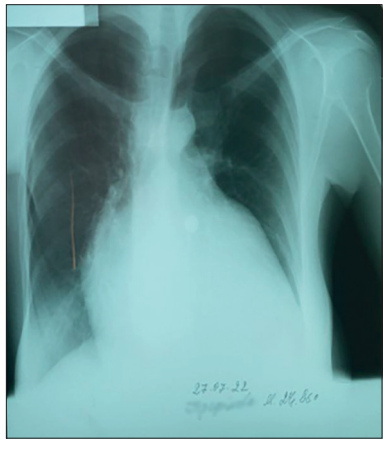

Fig. 3. *Chest X-ray before surgery (7/28/2022)*

+ tricuspid valve plasty with the imposition of a support ring + triangular plasty of the left atrium with its partial resection + resection of the right atrium+ resection of the LA appendage.

The duration of the operation was 350 minutes; it was performed under conditions of cardiopulmonary bypass (CPB) (159 minutes) and general hypothermia (27.8 °C). Aortic cross clamp time was 123 minutes. Intraoperative blood loss was 250.0 ml.

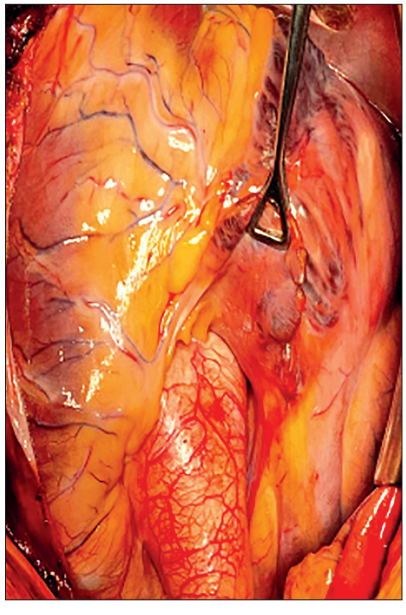

Fig 4. *The ascending aorta was 2.8-2.9 cm, and the sinuses of the Valsalva were in the range of 4.1-4.2 cm. Aortic tissues were not altered. Due to the significant severity of the patient's condition, preventive interventions at the aortic root were not provided, since the lesion of the aortic valve was minimal, the size of the ascending aorta was normal, and the tissues had certain structuring*

The operation was performed as follows. Median sternotomy. Minor dilatation of the sinuses of Valsalva (tissues were not altered) (Fig. 4). Aorta and vena cava cannulation. CPB. Antegrade cardioplegia in the aortic root with St. Thomas solution 400.0 ml, and later Custodiol retrograde solution 2300.0 ml. The dose of cardioplegia pumping was increased by 30% due to left ventriculomegalia. The duration of cardioplegia pumping was 25 minutes in total. The electrical activity of the heart disappeared on the 5th minute. Both atria were dissected separately.

Mitral valve revision: significant dilatation of the left atrioventricular foramen ring with significant myxomatous degeneration of the valves and their prolapse (Fig. 5) (end diastolic volume 373.0 ml). Valve plastic surgery is risky, so prosthetics are more reliable with maximum preservation of the valve space. The anterior leaflet of the mitral valve was plintered, the valve and subvalve structures were fully preserved (Fig. 6). Medtronic-31 mechanical prosthesis (in an intermediate position) was implanted on U-shaped separate sutures with gaskets in the amount of 14 pcs. The appendage of the left and right atria was cut off and sewn from the outside (Fig. 7). Triangular plasty of the LA was made (Fig. 8) using autopericardial strips to strengthen the barrage joints (Fig. 9). The use of an autopericardial strip significantly reduces blood loss. Additionally, resection of LA 5x2 cm along the incision line was performed (Fig. 10).

The tricuspid valve plasty with the imposition of Plankor-A support ring No. 30 was made. Resection of the right atrium along the incision line was performed. The heart resumed its activity through depolarization (once). Total warming of the patient to 36.8 °C was provided. After stopping the CPB: central venous pressure 60 mm of water, left atrial pressure 170 mm of water, blood pressure 110/70 mm Hg. Inotropic support after CPB stop: dobutamine at a dose of 2.5 μg/kg/min.

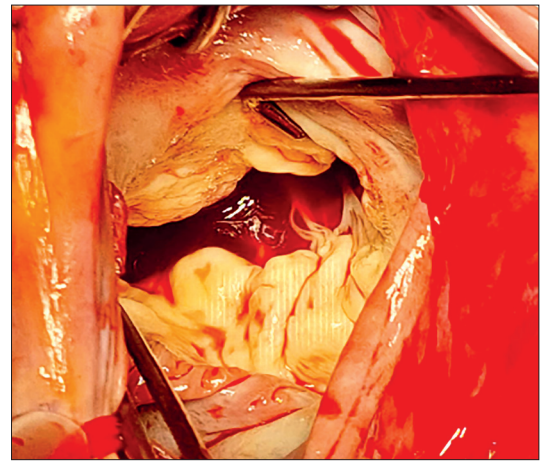

Fig. 5. *Significant dilatation of the ring of the left atrioventricular foramen with significant myxomatous degeneration of the valves and their prolapse*

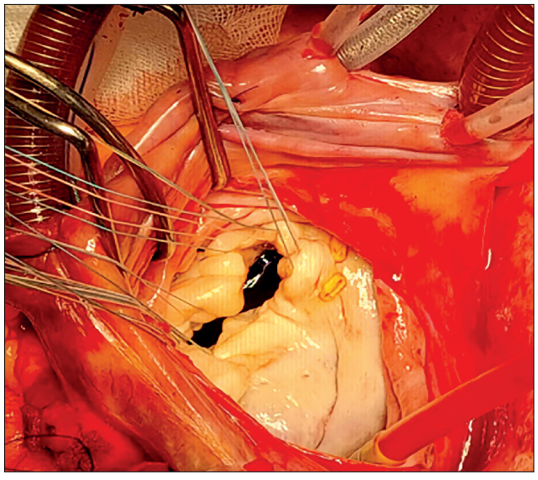

Fig. 6. *Complete preservation of the valves of the mitral valve during its replacement*

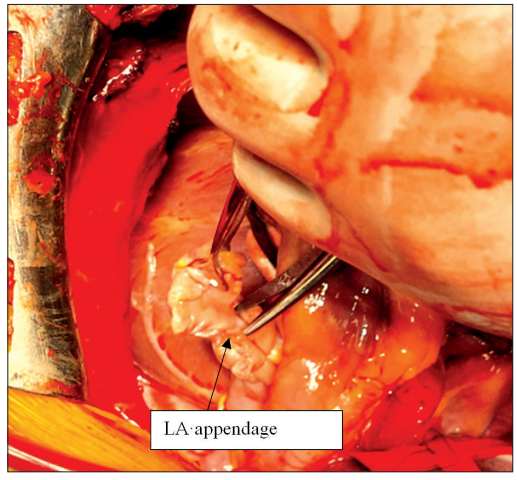

Fig. 7. *LA appendage resection*

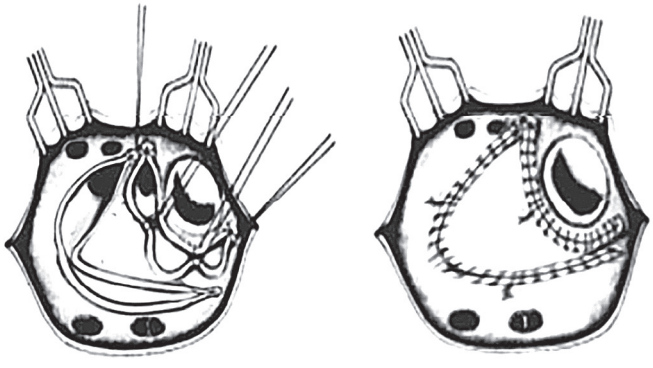

Fig. 8. *Scheme of triangular plasty of the LA developed by prof. Volodymyr V. Popov*

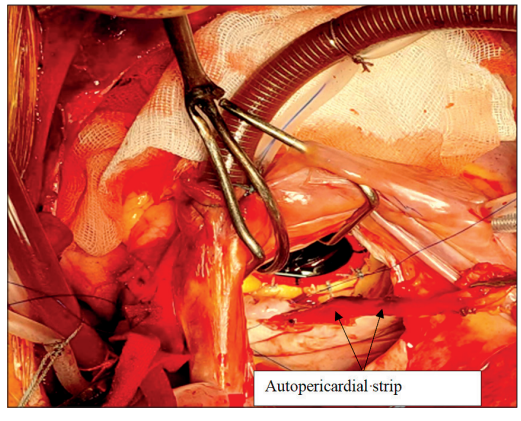

Fig. 9. *Use of autopericardial strip in triangular plasty of the LA*

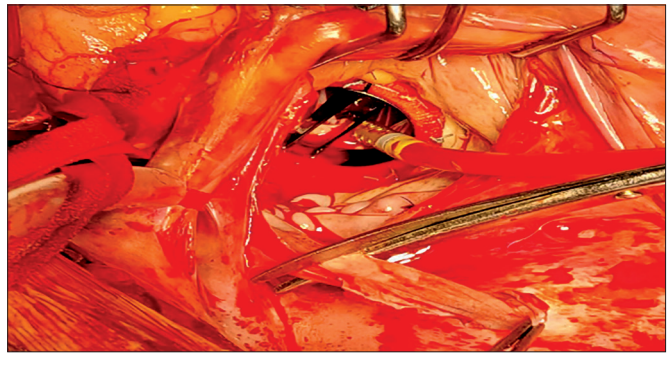

Fig. 10. *Additionally, resection of LA 5x2 cm along the incision line was performed*

The patient was extubated 4 hours after the end of the operation. The postoperative period in the intensive care unit lasted 96 hours. Inotropic dobutamine support on the first day after surgery was 1.3 μg/kg/min, on the second day it was stopped. Creatine kinase-MB level within 12 hours was 91 units/l. This indicates adequacy of myocardium protection during the operation.

While the patient was in the intensive care unit and received intensive care, asthenia, moderate bleeding were noted, and from day 3 the sinus rhythm was restored with an atrioventricular conduction of 0.14 ms, which indicates the adequacy of the correction. The NT-proBNP level, which was 51.6 times higher than the norm before surgery, was 8648 pg/ml on the 4th day after surgery, which is 1.3 times less than preoperative level but 39.3 times higher than normal, indicating the presence of latent HF that required intensive drug treatment.

The patient was transferred to the Department of Surgical Treatment of Acquired Heart Diseases, where drug therapy was successfully continued (Uperio, antibacterial, anticoagulant therapy, hormones, etc.).

Results. Table 1 demonstrates the echocardiographic parameters of the heart after surgery compared with preopera-

tive ones. There was evidence of a significant improvement in all indicators of cardiac morphometry (reduction of hemodynamic overload) by 52% on the 14th postoperative day.

The postoperative period was unremarkable. The wound healed by primary tension. Electrocardiography on discharge: AF with a frequency of 89/min. Chest X-ray demonstrated a decrease in heart shadow (Fig. 11).

The patient was discharged on 8/17/2022 (15 days after surgery) in satisfactory condition for further treatment in a cardiology hospital at the place of residence.

The patient underwent examination on 12/2/2022 (4 months after surgery) (Table 2).

According to Table 2, while 1 month after the operation the indicators were almost the same as those at discharge, within 4 months, when the phenomena of HF was substantially reduced due to intensive care (Uperio, prednisone), LVEF and systolic pulmonary artery pressure returned to normal.

Uperio has been used at a dose of 200 mg twice daily since discharge, which gave its positive results within 4 months, as evidenced by a decrease in NT-proBNP almost to normal and an increase in LVEF to normal.

NT-proBNP dynamics (pg/ml) (normal = 0–220.0) in the distant period:

9/3/2022 (1 month after surgery): 1822, which is 8.3 times higher than normal;

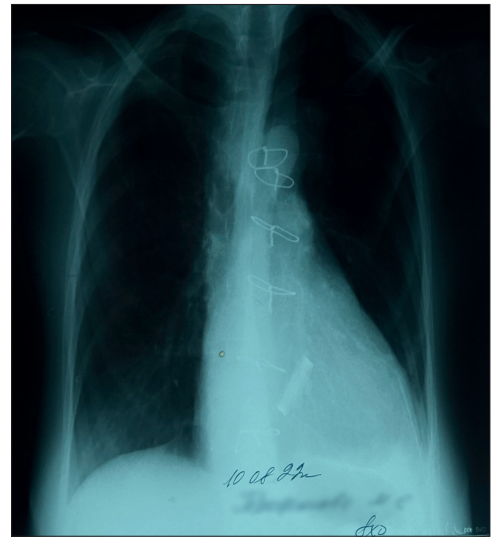

Fig. 11. *Chest X-ray after surgery (8/16/2022)*

10/11/2022 (2 months after surgery): 547, which is 2.5 times higher than normal;

12/1/2022 (4 months after surgery): 264, which is 1.2 times higher than normal.

At the time of the examination (12/2/2022), the patient's condition corresponded to the NYHA class II.

Table 2

Echocardiographic parameters in the distant period

Parameters (units)	Before the operation (7/27/2022)	1 month after surgery (9/8/2022)	4 months after surgery (12/2/2022)
Mitral valve	Myxomatous degeneration of the leaflets, prolapse of both valves, reverse leakage ++++	Prosthesis, reverse leakage minimal, pressure gradient 11 mm Hg	Prosthesis, reverse leakage minimal, pressure gradient 11 mm Hg
Tricuspid valve	Prolapse, reverse leakage expressed	Condition after tricuspid valve plasty, reverse leakage small, pressure gradient 1 mm Hg	Condition after tricuspid valve plasty, reverse leakage small, pressure gradient 6 mm Hg
Aortic valve	Tricuspid, compacted, Δp max 10 mm Hg, reverse leakage minimal, the diameter of the mouth of the aorta 2.2 cm, sinus of Valsalva diameter 3.9 cm, ascending aorta 3.0 cm	Tricuspid, compacted, Δp max 6 mm Hg, reverse leakage minimal, sinus of Valsalva diameter 4.2 cm, ascending aorta 3.1 cm	Tricuspid, compacted, Δp max 6 mm Hg, reverse leakage minimal, sinus of Valsalva diameter 4.1 cm, ascending aorta 3.4 cm
EDVI (ml/m^2)	233.1	120 (-48.5%)	133.1 (-42.9%)
LV EDV (ml)	373	192 (-48.5%)	213 (-42.9%)
ESVI (ml/m^2)	109.3	62.4 (-42.9%)	53.75 (-50.8%)
LV ESV (ml)	175	99.8 (-42.9%)	86 (-50.8%)
LV SVI (ml/m^2)	123.7	57.6 (-53.4%)	79.3 (-35.9%)
LV SV (ml)	198	92.2 (-53.4%)	127 (-35.9%)
LVEF	0.53	0.48 (-9.4%)	0.6 (+13.2%)
Diameter LA (cm)	7.2	4.9 (-31.9%)	4.5 (-37.5%)
SPAP (mm Hg)	40	38 (-5.0%)	32 (-20.0%)
RA diameter (cm)	6.5	3.9 (-40.0%)	4.0 (-38.5%)

She did not make any significant complaints. Previously diagnosed AF remained unresolved and required inotropic, arrhythmological support (digoxin, verapamil, and, from time to time, Uperio).

According to computed tomography dated 12/2/2022, there was no narrowing of the pulmonary veins after triangular plasty (Fig. 12).

Discussion. Marfan syndrome is a complex congenital pathology of single tissue which gives clinical manifestations at the level of valvular pathology and the ascending aorta.

When correcting a mitral-tricuspid defect in a patient with Marfan syndrome, the factor of biatriomegaly is clinically significant at the hospital stage, and to an even greater extent in the distant period, therefore, atrial reconstruction is an important component of treatment. The level of thromboembolic complications, as well as the level of cardiovascular insufficiency in the group without correction of left atriomegaly, in the distant period reaches a critical value, especially in combination with long-existing AF [1, 2, 3, 4]. Prophylactically, to reduce the likelihood of thromboembolic complications, resection of the LA appendage was performed, which is a mandatory procedure in the presence of AF. Triangular plasty of the LA should not lead to narrowing of the mouths of the pulmonary veins in the distant period.

Left atriomegaly in the presence of AF tachyform contributes to the occurrence of blood clots in the cavity of the LA and thromboembolic complications, therefore, during correction, it is necessary to make a maximum decrease in the volume of LA. In addition, dilated LA contributes to the progression of cardiorespiratory failure due to compression of the bronchi, trachea and posterior wall of the LV [5, 7, 8, 9, 10, 11].

Full restoration of the function of the right parts of the heart in Marfan syndrome leads (at the stage of 4 months after surgery) to the normalization of systolic pressure in the pulmonary artery and a decrease in the phenomena of latent heart failure (the level of NT-proBNP was almost normal within 4 months) [11, 12, 13].

It is important to note the discrepancy between the rate of LV remodeling and the decrease in the level of latent HF. As we observed at the time of discharge, the level of latent HF was still high and this required consideration in the long-term period (up to six months). Prolonged AF significantly affects the manifestation of HF and its tachyforms, which are difficult to stop, despite the sufficient digitalization of the patient.

This will be the subject of efforts by arrhythmologists in the long term, with possible endovascular isolation of the pulmonary veins throughout the year. For a young woman, the possibility of fertilization with subsequent caesarean section is an important factor in social rehabilitation.

In Marfan syndrome and mitral valve prosthetics, complete preservation of valvular and subvalvular structures

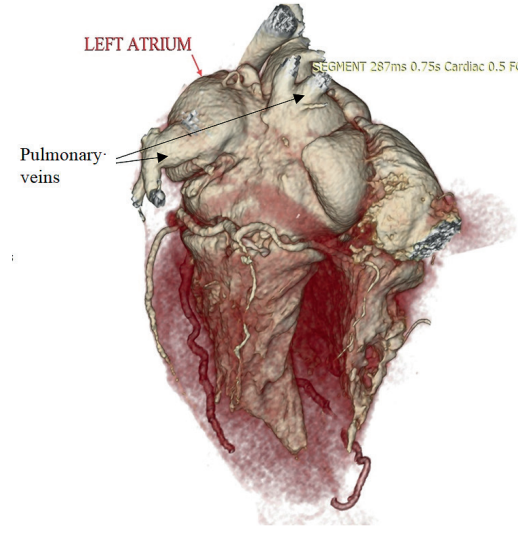

Fig. 12. *Computed tomography scan of the heart in the distant period. The structure of the mouths of the pulmonary veins after triangular plasty of the LA has not been changed*

can significantly reduce the critically increased volume of LV, maintain its contractility, especially in the presence of ventriculomegaly, which gives manifestations of critical HF [8, 9, 10, 11]. Functional recovery of the heart (decrease in the level of latent HF) is observed only by 4 months, and only thanks to powerful therapy (Uperio, prednisone), which is an urgent element of the treatment strategy, starting from the preoperative stage. Therefore, the legitimate question is that the drug Uperio is the gold standard for increasing the inotropic function of the heart, without having additional complications.

The most lethal complication of dilatation of ascending aorta and aortic arch is the development of aortic dissection or penetrating aortic ulcer. Average age of aortic dissection was found to be 36 ± 4 years [6].

The timing for aneurysm surgery is a balance between the risks imposed by surgical intervention and that of natural history of vascular pathology of the great vessels. The timing depends primarily on the diameter of the aneurysm, presence of risk factors for dissection of aorta, condition of the aortic valve, rate of growth of ascending aorta, aortic arch and experience of the surgical team.

The complexity of the decision by the surgeon on the advisability of prophylactic use of surgery on the ascending aorta runs counter to the clinical complexity of lesions of the left heart, as well as the presence of severe HF, which does not allow to increase the aortic cross clamp time and duration of CPB.

Conclusions. Surgical treatment of complex valvular pathology in Marfan syndrome without prophylactic correction of the ascending aorta is an important decision in the clinically complex course of the disease. An extremely neglected patient (NT-proBNP level is 51 times

higher than normal) with Marfan syndrome according to vital indications is eligible for surgical treatment by a surgical team with sufficient experience to significantly reduce the time of anoxia and maximize the radicality of the intervention. Surgical treatment of mitral-tricuspid defect with reduction of both atria is a desirable procedure in patients with biatriomegaly. Such an intervention leads to a significant improvement in morphometry, a decrease in hemodynamic overload of the heart, accompanied by a positive clinical effect on both hospital and remote periods. To achieve an effect of a significant reduction in HF, ideal myocardial protection is required, especially with left-wing ventriculomegaly and a long-standing (more than 10 years) permanent form of AF, which has happened and as evidenced by the very short period of inotropic support in the intensive care unit. Therefore, we consider the long-term retrograde supply of cardioplegic solution to be an ideal way to preserve myocardial reserves, taking into account the neglected condition of the patient (the level of NT-proBNP 51 times higher than normal).

In Marfan syndrome with the presence of left ventriculomegaly, mitral valve prosthetics should be performed with the maximum possible preservation of the sub-valve structures, the frame of the LV, in order to avoid any decrease in its contractility.

Thus, careful medical preoperative preparation, intensive treatment, adequate myocardial protection, comprehensive adequate reconstruction of the left and right parts of the heart during the correction of mitral-tricuspid defect can reduce the risks of complications of the hospital stage, even in extremely difficult patients. Unfortunately, the issue of long-standing AF, which will worsen the effect of the operation, limit functionality, remains unresolved. The effect of the operation in such a complex category of operated persons should be assessed only on the basis of the results of the first half of the year after the operation. The patient needs further close clinical supervision, given the possible deterioration of morphology on the aorta, which is natural in patients aged 35-39 years with Marfan syndrome.

Conflict of interest

The authors have no conflicts of interest to declare.

References

1. Kurfirst V, Mokráček A, Krupauerová M, Canádyová J, Bulava A, Pešl L, et al. Health-related quality of life after cardiac surgery--the effects of age, preoperative conditions and postoperative complications. J Cardiothorac Surg. 2014 Mar 11;9:46. https://doi.org/10.1186/1749-8090-9-46
2. Prabhu S, McLellan AJ, Walters TE, Sharma M, Voskoboinik A, Kistler PM. Atrial Structure and Function and its Implications for Current and Emerging Treatments for Atrial Fibrillation. Prog Cardiovasc Dis. 2015;58(2):152-167. https://doi.org/10.1016/j.pcad.2015.08.004
3. Abd Elaziz ME, Ibrahim IM. Reversal of Left Ventricular Functions in Chronic Mitral Regurgitation after Mitral Valve Replacement. Heart Surg Forum. 2016;19(4):E160-E164. https://doi.org/10.1532/hsf.1440
4. Chen L, Chen B, Hao J, Wang X, Ma R, Cheng W, et al. Complete Preservation of the Mitral Valve Apparatus during Mitral Valve Replacement for Rheumatic Mitral Regurgitation in Patients with an Enlarged Left Ventricular Chamber. Heart Surg Forum. 2013;16(3):E137-E143. https://doi.org/10.1532/HSF98.20121128
5. Guo Y, He S, Wang T, Chen Z, Shu Y. Comparison of modified total leaflet preservation, posterior leaflet preservation, and no leaflet preservation techniques in mitral valve replacement - a retrospective study. J Cardiothorac Surg. 2019 Jun 7;14(1):102. https://doi.org/10.1186/s13019-019-0918-7
6. Falk V, Baumgartner H, Bax JJ, De Bonis M, Hamm C, Holm PJ, et al.; ESC Scientific Document Group. 2017 ESC/EACTS Guidelines for the management of valvular heart disease. Eur J Cardiothorac Surg. 2017;52(4):616-664. https://doi.org/10.1093/ejcts/ezx324. Erratum in: Eur J Cardiothorac Surg. 2017 Oct 1;52(4):832.
7. Stassen J, Butcher SC, Namazi F, Ajmone Marsan N, Bax JJ, Delgado V. Left Atrial Deformation Imaging and Atrial Fibrillation in Patients with Rheumatic Mitral Stenosis. J Am Soc Echocardiogr. 2022;35(5):486-494.e2. https://doi.org/10.1016/j.echo.2021.12.010
8. Vohra S, Pradhan A, Jaiswal V, Sharma P, Pokhrel NB, Song D. Silent giant left atrium: A case report. Clin Case Rep. 2022;10(2):e05363. https://doi.org/10.1002/ccr3.5363
9. Liu QQ, Du Y, Zhang R, Shen WQ, Du GQ. Evaluation of longitudinal trajectory of functional tricuspid regurgitation on the risk of right ventricular dysfunction after mitral valve replacement. J Card Surg. 2022;37(11):3995-4001. https://doi.org/10.1111/jocs.16891
10. Suwanto D, Dewi IP, Budiarto M. Managing tricuspid valve pathology in multiple valvular heart disease. Ann Med Surg (Lond). 2022;82:104719. https://doi.org/10.1016/j.amsu.2022.104719
11. Collins N, Sugito S, Davies A, Boyle A, Sverdlov A, Attia J, et al. Prevalence and survival associated with pulmonary hypertension after mitral valve replacement: National echocardiography database of Australia study. Pulm Circ. 2022;12(4):e12140. https://doi.org/10.1002/pul2.12140
12. Yokoyama Y, Tsukagoshi J, Takagi H, Takayama H, Kuno T. Concomitant tricuspid annuloplasty in patients with mild to moderate tricuspid valve regurgitation undergoing mitral valve surgery: meta-analysis. J Cardiovasc Surg (Torino). 2022;63(5):624-631. https://doi.org/10.23736/S0021-9509.22.12354-2
13. Hasan IS, Qrareya M, Crestanello JA, Daly RC, Dearani JA, Lahr BD, et al. Impact of tricuspid valve regurgitation on intermediate outcomes of degenerative mitral valve surgery. J Thorac Cardiovasc Surg. 2022 Sep 26:S0022-5223(22)01025-X. Forthcoming 2022. https://doi.org/10.1016/j.jtcvs.2022.09.035

Хірургічна реконструкція мітрально-тристулкової недостатності в поєднанні з біатріомегалією при синдромі Марфана

Попов В. В.[1], д-р мед. наук, професор, завідувач відділу

Большак О. О.[1], канд. мед. наук, старший науковий співробітник

Букарім В. Ж.[1], молодший науковий співробітник

Хорошковата О. В.[1], канд. мед. наук, лікар-кардіолог

Гуртовенко О. М.[1], науковий співробітник

Новіков Є. В.[2], канд. мед. наук, асистент

[1]Відділ хірургічного лікування набутих вад серця, ДУ «Національний інститут серцево-судинної хірургії імені М. М. Амосова НАМН України», м. Київ, Україна

[2]Кафедра пропедевтики внутрішньої медицини, променевої діагностики та променевої терапії, Запорізький державний медичний університет, м. Запоріжжя, Україна

Резюме. Синдром Марфана – це комплекс серйозних дисфункцій тканин серця та магістральних судин, які становлять загрозу для пацієнта під час хірургічного лікування цієї патології. Хірургічне лікування мітрально-тристулкової вади з редукцією обох передсердь – бажана процедура у пацієнтів з біатріомегалією. Таке втручання призводить до значного покращення морфометрії, зменшення гемодинамічного перевантаження серця, супроводжується позитивним клінічним ефектом як на госпітальному етапі, так і у віддаленому періоді.

При синдромі Марфана з наявністю лівої вентрикуломегалії протезування мітрального клапана потрібно виконувати з максимально можливим збереженням підклапанних структур, каркасності лівого шлуночка, щоб уникнути будь-якого зниження його скоротливості.

Таким чином, ретельна медикаментозна доопераційна підготовка, інтенсивне лікування, адекватний захист міокарда, комплексна реконструкція лівих та правих відділів серця під час корекції мітрально-тристулкової вади дає змогу зменшити ризики ускладнень госпітального етапу навіть у вкрай важких пацієнтів. Ефект операції у такої складної категорії оперованих має бути оцінений тільки на підставі результатів віддаленого періоду після операції. Пацієнтові потрібний подальший щільний диспансерний нагляд, враховуючи можливе погіршення морфології на аорті, що є природним у пацієнтів віком 35–39 років при синдромі Марфана.

Мета – проаналізувати результати впровадження комплексної реконструкції лівих та правих відділів серця при синдромі Марфана в поєднанні з мітрально-тристулковою недостатністю, біатріомегалією та лівою вентрикуломегалією.

Матеріали та методи. Як приклад цієї патології ми представили таке спостереження. Хвора Т., 24 років, перебувала на обстеженні та лікуванні з 27.07.2022 по 17.08.2022 у відділі хірургічного лікування набутих вад серця ДУ «Національний інститут серцево-судинної хірургії імені М. М. Амосова НАМН України» з діагнозом: синдром Марфана, мітрально-тристулкова недостатність IV ст.; біатріомегалія; висока легенева гіпертензія; постійна форма фібриляції передсердь (з 2011 року – 10 років); серцева недостатність IIB з лівою вентрикуломегалією та зниженою фракцією викиду лівого шлуночка; функціональний клас за NYHA IV+. Хворій виконано оперативне втручання в об'ємі: протезування мітрального клапана з повним збереженням клапанного апарату, пластика тристулкового клапана з накладанням опорного кільця, трикутна пластика лівого передсердя з його частковою резекцією, резекція правого передсердя.

Результати. У терміни до 4 місяців після операції ехокардіографічні показники продемонстрували значне зменшення об'єму лівого шлуночка, діаметрів обох передсердь. Крім того, спостерігалося достовірне зниження рівня N-кінцевого пропептиду натрійуретичного гормону В типу (NT-proBNP), зменшення проявів серцевої недостатності.

Висновки. Враховуючи початковий тяжкий стан хворої із синдромом Марфана та вираженою мітрально-тристулковою вадою серця, лівою вентрикуломегалією зі зниженою фракцією викиду лівого шлуночка, біатріомегалією (ліве передсердя – 7,2 см і праве передсердя – 6,5 см), постійною формою фібриляції передсердь комплексна реконструкція лівих і правих відділів серця призводить до поліпшення функціонального стану міокарда. Прискіпливий диспансерний етап спостереження дасть змогу зменшити наслідки серцевої недостатності та тахіформи фібриляції передсердь.

***Ключові слова**: кардіохірургічні втручання, фібриляція передсердь, ліва вентрикуломегалія, штучний кровообіг, дилатація синусів Вальсальви.*

Стаття надійшла в редакцію / Received: 04.05.2023
Після доопрацювання / Revised: 15.06.2023
Прийнято до друку / Accepted: 24.06.2023